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Mitral valve repair during bypass in a 4-year-old girl with familial Evans syndrome

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In patients with Evans syndrome, autoantibodies of the IgG type are produced against red cells, platelets, and granulocytes, leading to phagocytosis of the coated cells by the reticuloendothelial system. This leads to recurrent episodes of intravascular hemolysis and thrombocytopenia in the affected individuals.[1] We report our experience of open mitral valve repair during cardiopulmonary bypass in a 4-year-old girl. This is the first reported case of a cardiac operation in a patient affected with this rare disorder.

CLINICAL SUMMARY

The patient presented in March 2004 with mild mitral regurgitation. She was known to have antibody-mediated hemolytic anemia and low platelets counts. This child came from a unique family in which 2 other siblings were given diagnoses of ''Evans syndrome.''

Her mitral valve regurgitation continued to worsen rapidly, leading to shortness of breath on mild exertion. Echocardiographic analysis in February 2005 showed severe mitral regurgitation with anterior mitral leaflet prolapse. Vena contracta was 6 mm, with an effective regurgitant orifice of 40% and a mildly dilated left ventricle with good systolic function.

Open mitral valve repair was carried out on December 2, 2006. A cardiopulmonary bypass machine with roller pumps was used during the operation. Moderate hypothermia (32°C), bicaval venous cannulation, and cold blood cardioplegic arrest were used. Usual heparinization (300 U/kg unfractionated heparin administered intravenously) with periodic activated clotting time measurement during bypass and reversal with protamine at the end of the procedure was performed. A routine cardiotomy sucker with return of the collected blood to a venous reservoir was also used.

The mitral valve had a dilated annulus and prolapse of the anterior mitral leaflet at the A2 segment, with elongated chordae.

Chordal repositioning of the involved segment was carried out. The corresponding papillary muscle was split longitudinally, and the tip containing the elongated chordae was bent downward on itself. This accomplished shortening of the chordae to the A2 segment. The tip of the repositioned (bent downward) half of the papillary muscle was sutured to the other half of the papillary muscle with 5-0 Prolene sutures. Edge-to-edge modified leaflet placation with 5-0 Tycron sutures was done on both commissures. A 27-mm St Jude Medical Tailor ring annuloplasty was carried out. Intraoperative transesophageal echocardiographic analysis confirmed only trace mitral regurgitation at the posteromedial commissure.

On the fourth postoperative day, dark (brown)–colored urine was noticed, with a significant decrease in hemoglobin levels. Analysis of urine confirmed hemoglobinuria. Peripheral blood smear microscopy showed scant fragmented red cells. The reticulocyte counts and serum lactate dehydrogenase levels were increased. A trial of intravenous immunoglobulin and steroids did not completely resolve the hemolysis.[1] The patient required blood transfusion twice per week. She was discharged home 3 weeks after the operation.

From a cardiovascular point of view, she did well. Repeat echocardiographic analysis confirmed mild mitral regurgitation, a small regurgitant jet at the posteromedial commissure, and good biventricular function. The regurgitant jet was considered not significant on echocardiographic analysis.

She was followed up by the pediatric hematology section of our hospital. The girl required frequent admissions in the following months during which she received blood

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transfusions on a weekly basis to correct her anemia. Complete courses of cyclophosphamide and rituximab also failed to resolve the hemolysis and subsequent anemia.

In the subsequent months, the spleen became progressively enlarged. All the clinical and laboratory data indicated increased intravascular hemolysis. It was decided to perform a splenectomy in this child. However, she became septic and died of multiorgan failure 8 months after mitral valve surgery during one of her hospital admissions before splenectomy.

DISCUSSION

We did not see excessive bleeding, hemolysis, coagulopathy, or thrombosis during or immediately after cardiopulmonary bypass.

In subsequent weeks, after her mitral valve repair, persistent hemolysis became troublesome. High shear stresses caused by flow disturbances in some cases with mitral valve repair lead to significant hemolysis. The culprit regurgitant jets have identifiable echocardiographic patterns.[2]

Evans syndrome manifests as recurrent episodes of intravascular hemolysis with spontaneous or therapy-induced remissions.[3] Immune-mediated intravascular hemolysis also leads to the presence of fragmented red cells in peripheral blood smears, increased reticulocytes counts, and an increase in plasma free hemoglobin and lactate dehydrogenase levels.[4]

Persistence and increased severity of hemolysis in this child after her mitral valve repair prompts us to believe a role was played by mechanical hemolysis after mitral valve surgery. Uncomplicated bypass in patients with nocturnal hemoglobinuria is reported.[5]

With the benefit of hindsight, one should take into account the limited lifespan of patients with Evans syndrome as a factor in decision making for cardiac surgery. It might also be prudent to use measures to reduce hemolysis during bypass, such as use of centrifugal pumps, separation of cardiotomy blood, avoidance of suction-assisted venous drainage, minimal use of intracardiac prosthetic materials, and short bypass time, if possible.

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Ruptured aneurysm of the sinus of Valsalva protruding into the pulmonary artery

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Congenital sinus of Valsalva aneurysm is uncommon. The rupture results in a fistulous connection between the aorta and any cardiac chamber. This report describes a patient with a ruptured aneurysm of the right sinus of Valsalva that extended to the right ventricle, crossed over the pulmonary valve, and protruded into the main pulmonary artery. The aneurysm obstructed the right ventricular outflow tract (RVOT) and made a pressure gradient between the right ventricle and the pulmonary artery. The top of the aneurysmal sac had a ruptured hole opening in the pulmonary artery that caused an aortopulmonary shunt. To the best of our knowledge, no other cases with these features have been reported in the medical literature.

CLINICAL SUMMARY

A 46-year-old man presented with dyspnea on exertion, which he had experienced for the past several years. A heart murmur had been diagnosed in the patient at the age of 7 years. A ventricular septal defect was suspected, but the patient had not undergone a checkup because he had no symptoms. On presentation, he was in New York Heart Association class II.

Transthoracic echocardiography revealed an aneurysm of the right sinus of Valsalva that extended into the RVOT. An

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